Complete Summary

GUIDELINE TITLE

Focal segmental glomerulosclerosis: use of other therapies.

BIBLIOGRAPHIC SOURCE(S)

Thomas M. Focal segmental glomerulosclerosis: use of other therapies. Nephrology 2006 Apr;11(S1):S196-7.

Thomas M. Focal segmental glomerulosclerosis: use of other therapies. Westmead NSW (Australia): CARI - Caring for Australasians with Renal Impairment; 2005 Sep. 4 p. [8 references]

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Focal segmental glomerulosclerosis
- End-stage kidney disease

GUIDELINE CATEGORY

Management
Treatment

CLINICAL SPECIALTY

Family Practice
Internal Medicine
Nephrology
Pediatrics

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To evaluate the available clinical evidence pertaining to the impact of interventions not covered in other guidelines on renal functional decline in patients with idiopathic focal segmental glomerulosclerosis

TARGET POPULATION

Adults and children with idiopathic focal segmental glomerulosclerosis resistant to standard treatment

INTERVENTIONS AND PRACTICES CONSIDERED

Treatment*

1. Mycophenolate mofetil
2. Tacrolimus
3. Vincristine
4. Plasma exchange
5. Lipid apheresis
6. Immunoadsorption

*Considered but not recommended

MAJOR OUTCOMES CONSIDERED

- Improvement in proteinuria
- Remission of focal segmental glomerulosclerosis

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Databases searched: MeSH terms and text words for focal segmental glomerulosclerosis were combined with MeSH terms and text words for mycophenolate mofetil, tacrolimus, vincristine, plasma exchange, lipid apheresis, immunoadsorption and other therapies. This search was carried out in Medline

(1966 to September Week 2, 2004). The Cochrane Renal Group Trials Register was also searched for trials not indexed in Medline.

Date of searches: 17 September 2004.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

Level I: Evidence obtained from a systematic review of all relevant randomized controlled trials (RCTs)

Level II: Evidence obtained from at least one properly designed RCT

Level III: Evidence obtained from well-designed pseudo-randomized controlled trials (alternate allocation or some other method); comparative studies with concurrent controls and allocation not randomized, cohort studies, case-control studies, interrupted time series with a control group; comparative studies with historical control, two or more single arm studies, interrupted time series without a parallel control group

Level IV: Evidence obtained from case series, either post-test or pretest/post-test

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Comparison with Guidelines from Other Groups
Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Recommendations of Others. Recommendations regarding blood pressure control targets in chronic kidney disease from the following groups were discussed: Kidney Disease Outcomes Quality Initiative, UK Renal Association, Canadian Society of Nephrology, European Best Practice Guidelines, and International Guidelines.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Definitions for the levels of evidence (I–IV) can be found at the end of the "Major Recommendations" field.

Guidelines

No recommendations possible based on Level I or II evidence

Suggestions for Clinical Care

(Suggestions are based on Level III and IV sources)

There have been a number of case series using mycophenolate mofetil in patients with resistant focal segmental glomerulosclerosis (FSGS). Most demonstrate that although mycophenolate mofetil can induce some reduction of proteinuria, complete remission of proteinuria is rare. No data on long-term follow-up evaluation with this drug are currently available.

- One group of authors performed an open-label, 6-month trial of mycophenolate mofetil in 18 patients with biopsy-proven FSGS who were resistant to corticosteroid therapy. Seventy-five percent had also failed to respond to a cytotoxic agent and/or a cyclosporin. A substantial improvement in proteinuria was seen in 44% (8/18) of patients by 6 months. However, no patient achieved complete remission. In addition, relapses were common after therapy was discontinued.

- One group of authors previously reported the use of mycophenolate mofetil in 7 patients, in whom a substantial improvement in proteinuria was also observed.
- One group of authors investigated the effect of mycophenolate mofetil in 7 children with a resistant nephrotic syndrome (6 of whom had minimal change disease and one with FSGS). In this patient, mycophenolate mofetil resulted in complete remission for a follow-up of 28 months.

Other therapies have been used in patients with FSGS who prove resistant to standard treatment:

- Partial remission has been observed in a few case reports using tacrolimus.
- Vincristine has also been used for the treatment of steroid- and cyclophosphamide-resistant nephrotic syndrome. In a series of eight cases presented by one group of authors, two children treated with vincristine achieved complete remission associated with preserved renal function. Another experienced transient relapses. Although studied in primary FSGS, there may be particular advantages of vincristine in secondary forms of nephrotic syndrome associated with malignancy (see Guideline titled "FSGS: cytotoxic therapy" in the original guideline document).
- Plasma exchange, lipid apheresis and immunoadsorption have also been reported to induce remission of proteinuria in selected patients.

Definitions:

Levels of Evidence

Level I: Evidence obtained from a systematic review of all relevant randomized controlled trials (RCTs)

Level II: Evidence obtained from at least one properly designed RCT

Level III: Evidence obtained from well-designed pseudo-randomized controlled trials (alternate allocation or some other method); comparative studies with concurrent controls and allocation not randomized, cohort studies, case-control studies, interrupted time series with a control group; comparative studies with historical control, two or more single arm studies, interrupted time series without a parallel control group

Level IV: Evidence obtained from case series, either post-test or pretest/post-test

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate management of patients with focal segmental glomerulosclerosis (FSGS)
- Remission of FSGS

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2006 Apr

GUIDELINE DEVELOPER(S)

Caring for Australasians with Renal Impairment - Disease Specific Society

SOURCE(S) OF FUNDING

Industry-sponsored funding administered through Kidney Health Australia

GUIDELINE COMMITTEE

Not stated

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Author: Merlin Thomas

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All guideline writers are required to fill out a declaration of conflict of interest.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the Caring for Australasians with Renal Impairment Web site.

Print copies: Available from Caring for Australasians with Renal Impairment, Locked Bag 4001, Centre for Kidney Research, Westmead NSW, Australia 2145

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- The CARI guidelines. A guide for writers. Caring for Australasians with Renal Impairment. 2006 May. 6 p.

Electronic copies: Available from the Caring for Australasians with Renal Impairment (CARI) Web site.

PATIENT RESOURCES

None available

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